

CLINICAL AND HISTOLOGICAL STUDY OF A MELANOTIC
SARCOMA OF THE CHOROID, WITH RECURRENCE
OF THE GROWTH IN THE ORBIT FIVE MONTHS
AFTER ENUCLEATION OF THE EYEBALL
AND ONE EXTRA-SCLERAL MASS.*

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PHILADELPHIA.

Illustrated.

Certain interesting features attach themselves to the clinical history of this case of sarcoma of the choroid, which we desire to relate in connection with a description of the macroscopic and microscopic specimens.

Herman S., aged 36, a butcher, presented himself for treatment to one of us (Dr. de Schweinitz) on the 10th of May, 1899.

History.—There is nothing of importance in the patient's general clinical history, nor in that of his family. At the age of six he acquired convergent strabismus, and the defective vision associated with that condition was noted. There is no history of injury to his eye and none of morbid growth in the patient's immediate family. Five years ago he consulted an ophthalmic surgeon, and was told that there was a growth in his left eye and that the eyeball ought to be enucleated. He declined to follow this advice, and also similar advice given about three years later, *i. e.*, on the 14th of August, 1897. It is probable that at both of these examinations the media were still clear, and that the growth was visible to the ophthalmoscope. He was driven to seek help in May, 1899, on account of severe pain, which had been growing worse during the past year.

Examination.—The vision of the right eye, after the correction of a high hypermetropia, was 6/9. The optic disc was congested. There was faint perivascularitis and slight superficial choroidal disturbance.

The vision of the left eye was *nil*; the bulbus was prominent, that is, slight exophthalmos was present; there was no bruit and no limitation in the movement of the eyeball. The iris was congested and adherent to the lens, which was cataractous. Large swollen episcleral vessels crossed the globe, especially at the inner side, and the ciliary zone was intensely injected. The tension was +2.

*Specimens presented to the Pathological Society of Philadelphia December 14, 1899.

The patient was informed that enucleation was imperative, the diagnosis of intraocular growth in the glaucomatous stage being only too evident, while the exophthalmos indicated the presence of extra-scleral nodules.

On May 12, 1899, the eyeball and one large densely pigmented mass which was imbedded in the upper portion of the orbit were enucleated at the Jefferson Medical College Hospital. These growths were placed in a 5 per cent solution of formaldehyde, and after hardening divided, one-half being reserved for macroscopic and the other for microscopic examination.

The patient made a rapid recovery, leaving the hospital on the fifth day after operation, and a month later was fitted with an artificial eye. The orbit continued to be perfectly healthy until the early portion of September, when a slight swelling was noted in its upper and inner portion, causing some œdema of the surrounding tissues. This swelling gradually increased, and a month later had developed into a large mass, which so filled the upper part of the orbit that it was not possible longer to insert the artificial eye. The patient was advised to submit to immediate exenteration of the orbital contents, but could not make up his mind to permit this operation until three weeks later, at which time the orbital cavity was packed with the secondary nodules and the patient suffering violent pain in the head.

On the 27th of October, 1899, the entire contents of the orbit were enucleated, together with the periosteum, in a single mass, which was composed almost entirely of a collection of densely pigmented nodules. It was found that the floor of the orbit had been eroded so that the antrum was open, but itself not yet involved by the growth, while on the inner side the ethmoid region had been perforated, and communicated in a softened track with the sphenoid. All suspicious tissue was removed, and the cavity packed with iodoform gauze. The patient, with the exception of slight constitutional disturbances dependent upon the absorption of iodoform, made an uninterrupted recovery, leaving the hospital on the thirteenth day. Thus far there has been no return of the growth, although it is too much to suppose that this will not sooner or later occur.

Macroscopic Examination.—The eye measured 25 mm. in its vertical and 23 mm. in its horizontal diameter. The neoplasm is seen to occupy the posterior third of the eyeball in the form of a flattened growth extending 15 mm. to the right of the optic nerve entrance and 13 mm. to its left, the width of the growth being respectively 4 and 3 mm. In other



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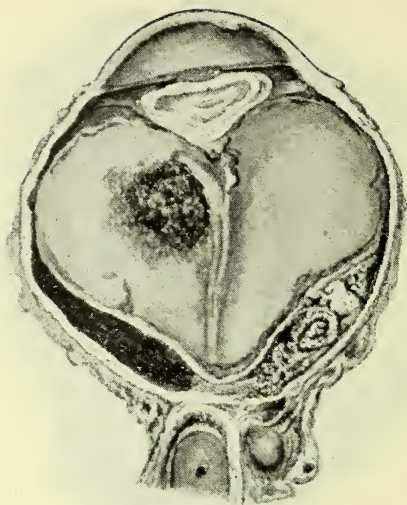


FIG. I. Macroscopic appearance of the choroidal sarcoma—flattened growth or so-called cake-like form.

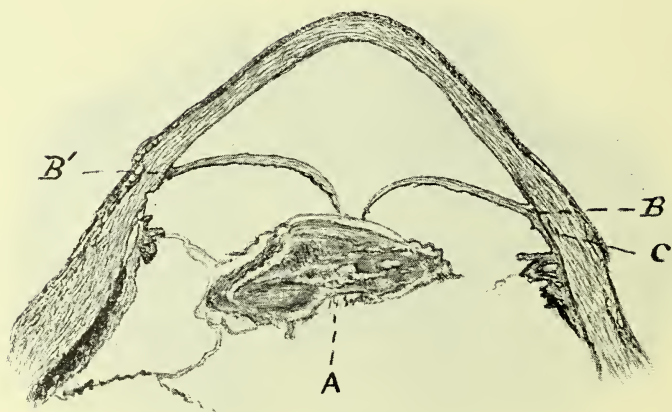


FIG. II. Occlusion of the angle of the anterior chamber by adhesive inflammation of iris base, B and B'; Schlemm's Canal, C.; remains of cataractous lens, and the adherent iris, A.

words, the growth springs from the choroid, and is somewhat circular in arrangement, surrounding the optic nerve entrance, but not approaching quite to it; that is, instead of the ordinary knob-like or spheroidal form, the growth has assumed this flattened, or, as it has been called by Mr. A. Hill Griffith, cake-like shape. One detached nodule, just posterior to the lens in the anterior portion of the vitreous, 6 by 5 mm., is evident. The part of the growth to the right of the optic nerve is mixed white and black, the section being not unlike Italian marble, while that upon the left side, as well as the separated portion, is densely brown-black in color. The retina is totally detached, and extends in a narrow band from the optic nerve entrance to the posterior portion of the lens. The lens itself is partially cataractous, and to it is attached completely the thickened and infiltrated iris. The cut surface of the optic nerve is slightly yellowish in color, and near its center contains a small pigment spot. On each side of the optic nerve are extra-scleral nodules, that upon the right, closely in contact with the nerve at the posterior surface of the eyeball, being 1 cm. in length and 5 mm. in width, while that upon the left is 4 mm. in length and 2 mm. in width. (Fig. I.)

The nodule found in the upper portion of the orbit at the time of the primary enucleation was a densely brownish-black mass, 2 cm. in length and 18 mm. in width, and only slightly, or practically not at all, adherent to the surrounding orbital tissue. The recurrent growth was a mass the size of the cerebellum, and not unlike it in shape, was closely invested by the periosteum, and on section was seen to be composed of three lobules, separated by fibrous trabeculæ. The main body of the tissue is of a densely brown-black color, and evidently repeats in structure the smaller nodule previously described.

Microscopical Examination.—The eye was imbedded in celloidin, and sections were cut and stained with Delafield's hematoxylin and eosin. The *cornea*, in so far as its anterior epithelium, anterior elastic layer, and substantia propria are concerned, is normal. The posterior elastic layer is somewhat uneven and detached from the cornea, while the endothelial layer lining the anterior chamber is thickened, with proliferation of its cells.

As the epithelium from the cornea passes over the corneo-scleral junction it becomes very much increased in thickness, and beneath it the veins are greatly distended and surrounded by a brightly-staining, small-celled infiltrate, evidently the representatives of the distended blood vessels which were noted crossing the bulbus prior to the enucleation.

The *iris* is typically bombé, thickened and infiltrated with small cells, pigment cells, and pigment granules, and is attached in a complete posterior synechia to the capsule of the lens, the attachment itself being composed of a similar mixture of small cells, pigment grains, and pigmented cells. The iris-base is closely adherent to the angle of the chamber, which is absolutely occluded, affording thus a typical example of the adhesive inflammation in this region, which prevents filtration and causes the glaucomatous stage of intra-ocular growths. (Fig. II.)

The *ciliary body* and *ciliary processes* are flattened and atrophied, and on the right side between the ciliary body and the pars ciliaris retinae is a narrow, spindle-shaped band of densely pigmented neoplastic tissue composed of cells like those presently to be described. The narrowed end of this deposit gradually merges into a strip of unaffected choroid, beyond which the second focus of sarcoma gradually develops in a large spindle-shaped area, reaching almost to the optic nerve entrance.

The *growth* is very deeply pigmented by brownish granular pigment, some of which is contained in the cells and some in the intercellular substance. The cells of the tumor are mostly small spindle-shaped ones with oval nuclei, but there is also a considerable number of large spindle-shaped and round cells, with large deeply staining round nuclei, and also smaller round cells with similar nuclei. These cells are seen especially about the smaller arterioles, where they are massed together about the vessel and present the appearance of a perivascular sarcoma. From their position and appearance it appears most probable that they have arisen from the perithelium of the artery. It is only the arteries with true walls that have these surrounding masses of cells. There is a delicate intercellular substance between the cells that stains faintly with eosin, and a very delicate reticulum of connective tissue running through the tumor, but in no place does this attain an amount sufficiently large to be considered a stroma. The blood vessels are fairly numerous. In many places they lack true walls, and are contained in a single layer of endothelium. In other places they have true walls, and are surrounded by the cylinder of cells already noted. In none of the sections do the walls of the vessels appear to be infiltrated by the growth. (Fig. III.)

Upon the opposite side there is the same adhesive inflammation of the base of the iris and occlusion of the anterior chamber, with atrophy of the ciliary body and ciliary processes, but no neoplastic deposit in connection with the ciliary body. Further on begins a similar flattened mass of sarcoma, less pigmented than upon the opposite side.

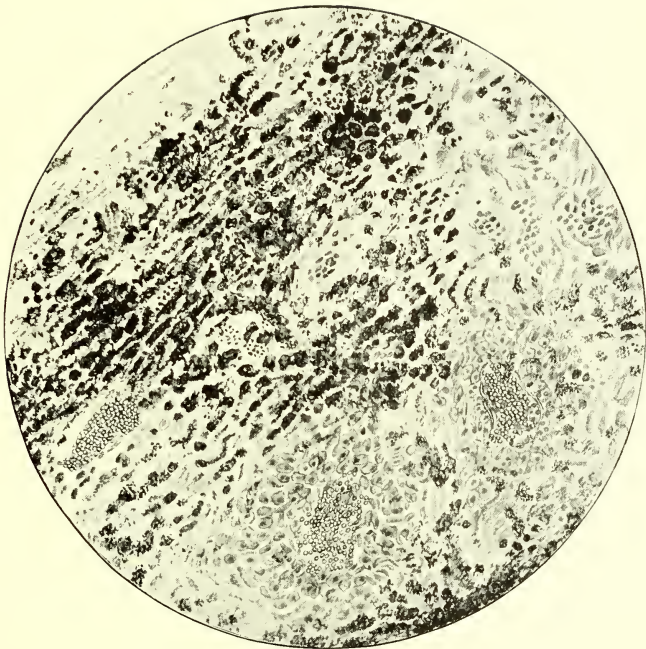


FIG. III. General microscopic appearance of the growth, showing pigmented cells, non-pigmented cells surrounding blood-vessels without walls, or with only a layer of endothelium surrounding the blood stream. Arteries with true walls do not appear in this section. (Zeiss' Ocular 8, obj. 16 mm.)



FIG. IV. Section of the optic nerve showing infiltration with sarcoma-cells, A, and central artery of the retina, B, filled with pigment granules and pigment bearing cells. Some of the cells appear black in the reproduction, as if they were pigmented; only those cells inside of the vessel were pigmented. (Zeiss' Ocular 8, obj. 16 mm.)

The description already given of the cellular elements, intercellular stroma and blood vessels which constitute the growth applies equally well to this portion of the neoplasm, which differs from the other only in the smaller quantity of pigment.

In the *vitreous chamber* are vitreous shreds, pigment granules, round and oval pigment cells, and numerous blood corpuscles caught in fibrinous prolongations.

The sections do not show the *optic entrance* nor the *retina*, which, as already described, was completely detached, and is contained in the half of the eyeball devoted to macroscopic study.

The fibrous septa separating the nerve bundles of the *optic nerve* are markedly thickened. There appears to be no change in the optic nerve so far as its nerve elements are concerned, but its connective tissue framework is smartly infiltrated with numerous round cells, with large round nuclei and comparatively little protoplasm, which are identical morphologically with the tumor cells. These cells do not occur in masses, but are scattered quite regularly and thickly through the fibrous tissue of the nerve. They are not representatives of the ordinary increase of the neuroglia nuclei, or of the nuclei of the interfascicular connective tissue, but may be regarded as neoplastic elements, or, in other words, there is a sarcomatous infiltration of the nerve. Precisely similar cells in much greater quantity fill the intersheath and the arachnoidian spaces. The growth in the choroid immediately adjacent to the nerve is not as considerable as it is farther out. In none of the sections examined can a connection be traced from the tumor to the optic nerve. (Fig. IV. A.)

The *central artery of the retina*, which is well seen in cross-section, is completely filled with a brownish homogeneous substance in which are imbedded numerous variously shaped and sized pigment grains and many round cells packed with pigment granules. Exactly similar pigment-bearing plugs and cells are visible in some of the smaller vessels of the optic nerve sheath and of the tissue adherent to the sclera. Elsewhere among the blood corpuscles contained in many of the arterioles are cells of medium size, and usually round, that contain the previously described brown pigment, evidently of the same character as that in the tumor. As before stated, the walls of these vessels are not infiltrated by the growth. The fact that these cells usually have irregular nuclei and occur singly and not in clumps makes it probable that they are leucocytes or mastzellen which have taken up the pigment, and that they are not sarcoma cells. Where this taking up of the pigment occurred does not appear. (Fig. IV. B.)

